Case Report

Indian Journal of Cancer Education and Research 77
Volume 9 Number 2, July-December 2021
DOI: http://dx.doi.org/10.21088/ijcer.2321.9815.9221.3

A Rare Case Report of Metastatic Alveolar Soft Part Orbital Sarcoma

Vividha Dubey[1], Jayeeta Sen[2], Saurabh Karnawat[3], Virendra Bhandari[4]

Author's Affiliation: [1,2]Registrar, [3]Assistant Professor, [4]Professor & Head, Department of Radiation Oncology, Sri Aurobindo Medical College & PG Institute, Indore 452001 Madhya Pradesh, India.

Corresponding Author: Virendra Bhandari, Professor & Head, Department of Radiation Oncology, Sri Aurobindo Medical College & PG Institute, Indore 452001, Madhya Pradesh, India.

E-mail: virencancer@yahoo.co.in

How to cite this article:

Vividha Dubey, Jayeeta Sen, Saurabh Karnawat et al./ A Rare Case Report of Metastatic Alveolar Soft Part Orbital Sarcoma/ Indian J Canc Educ Res 2021;9(2):77-80*.

Abstract

Alveolar soft part sarcoma (ASPS) is a morphologically distinct soft tissue tumour which is rare in its incidence. Most commonly it occurs in extremities in adults. Incidence as orbital tumour is rare and it relapses easily. Owing to its aggressive nature it requires early implementation of management with surgical, radiotherapy and chemotherapy as combined modality.

Keywords: Alveolar soft part sarcoma; orbital tumour.

Introduction

Alveolar soft part sarcoma (ASPS) is a rare malignant soft tissue lesion typically seen in younger adolescent individuals, especially those between 15 and 35 years of age. It is rare in children under 5 or in adults over 50. Women outnumber men, especially under age 25. There appears to be no link of this tumour to a particular ethnicity. ASPS accounts for about 0.5-1% of all soft tissue sarcomas.[1] The name "alveolar" was derived from its pseudo alveolar appearance with clustered polygonal cells lacking central cohesion. It is interesting to observe, that, these tumour cells exhibit characteristic PAS-positive, diastase resistant, intra-cytoplasmic rhomboid crystals that contain monocarboxylate transporter 1 and CD147.[2] Recent advanced molecular studies identified chromosome rearrangement der(17) t(X;17)(p11;q25) resulting in the ASPL-TFE3 fusion gene, which is highly specific and critical for development of the tumour.[1,3] In spite of these advances in tumour molecular biology research, the origin/differentiation of ASPS still remains uncertain and no standard effective treatment has been devised, especially for cases with advanced presentation.

Further rarity of the disease is additive to difficulties in understanding of the clinical behaviour and optimal treatment of ASPS. The majority of previous reports have been in the form of small collective series from different small referral facilities or multi-institutional studies over a long period suggested that ASPS is resistant to conventional cytotoxic chemotherapy and support the contention that complete excision of the tumour is the only meaningful treatment for ASPS. Moreover, recent data explore the clinical value of radiotherapy and cancer immunotherapy for treatment of ASPS.[4-14]

Here we present a case of 33 yrs male, known case of sarcoma, presented with multiple metastasis diagnosed on biopsy and immune-histochemical confirmation as alveolar soft part sarcoma. We share our experience of treatment approach through radiotherapy of this rare chemo resistant tumour.

Case report

A 30 yrs old male, known case of orbital sarcoma Post Radiotherapy in 2014 on adjuvant treatment and regular follow-up. In 2018 he presented with complaints of headache followed by recurrent vomiting multiple episodes and generalised weakness. CT brain revealed of brain metastasis.

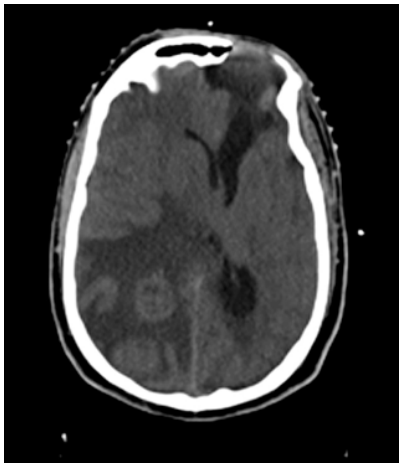

Fig. 2: CT scan showing lesion in brain S/O brain metastasis.

He received 30Gy/10# palliative RT to brain. USG whole abdomen revealed mild hepatomegaly with lesion in right lobe of liver measures approx 5.8 x 5.4 cm size with thick irregular wall associated with perilesional edema and central cystic component. CECT abdomen.

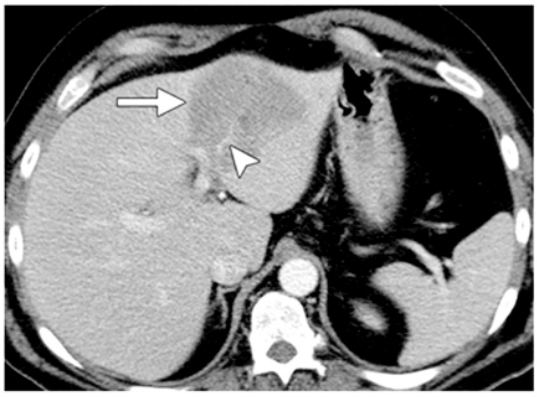

Fig. 3: CECT abdomen showing heterogeneous enhancing central necrotic lesion in right lobe of liver.

revealed sign of moderately enlarged liver and peripheral heterogeneous enhancing central necrotic likely neoplastic mitotic lesion in right lobe of liver measures approx. 5.2x4.8x4.6 cm size with prominent geographical enhancement in left lobe segment IVA with few density heterogeneously enhancing metastatic nodular lesion in bilateral lung parenchyma, largest in right lower lobe 3.6x3.2 cm. CT guided liver biopsy showed well differentiated adenocarcinoma (?Primary/metastatic). On imm -unohistochemistry, tumour cells are positive for TTF3 while negative for Hepar1, Glypican 3, synaptophysin and chromogranin. Final diagnosis of metastatic deposits of alveolar soft part sarcoma.

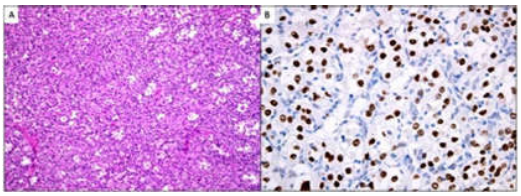

Fig. 1: A. Alveolar soft part sarcoma, solid type: large round and polygonal cells with abundant eosinophilic granular cytoplasm (H&E, Low power) B. Alveolar soft part sarcoma: positive nuclear staining for TFE3 (high power).

He was started on Tablet Pazopanib in 2018 and was on regular follow up. In December 2020 WBPET-CT scan revealed 2.4X2.4cm lesion Rt. Lung, and 3.3X3.3 cm lesion in liver and also splenic metastasis. Patient was then started on Bevacizumab and Sorafenib and put on regular follow-up. He however developed disease progression and succumbed in march 2021.

Discussion

ASPS is an extremely rare tumour of young adolescents and this makes it difficult to draw definitive conclusions regarding its clinical characteristics, prognostic factors, and appropriate treatment. The majority of previous case reports are form secondary referral centres or small collective series.[4–14] Here we share experience at tertiary advance centre in the era of modern multidisciplinary treatment[8,11,12] to uncover a treatment strategy and understand the prognosis of this rare tumour.

ASPS accounts for 0.5 - 1.0% of all soft tissue tumours. With most commonly arising in deep soft tissue of oral cavity, pharynx, mediastinum and thigh / leg. It is usually more common in young females. This tumour has highly malignant behaviour, although clinical course is slow / indolent and metastases can be seen as late as 30 years to lungs, liver, brain, orbit and other sites. Lung metastases may be presenting feature independently. Our case presented with multiple metastasis. Metastasis can be observed with CT scan. Histopathological examination reveals well defined nests of cells separated by fibrous stroma, alveolar pattern of discohesive cells, composed of large polygonal cells with granular eosinophilic cytoplasm, vesicular nuclei, prominent nucleoli. Vascular invasion is common and also characteristic rod-shaped

crystalloids. There are no/rare mitotic figures and minimal pleomorphism. Immunohistochemistry shows TFE3 nuclear positivity in tumour cells that is suggestive of characteristic translocation. (Figure 1) These cases are negative for EMA, Cytokeratins, HMB45, Melan-A, Chromogranin A and Synaptophysin.

Local recurrence after excision of ASPS was reportedly as high as 20% in the largest series of 91 patients treated at the Memorial Sloan-Kettering Cancer Center[8]. Evans noted a higher local recurrence rate of 31%.[13] In contrast, Portera et al. and van Ruth et al. reported that the local recurrence rate of ASPS after surgical excision was around 10%[11, 14] The risk factors for local recurrence in ASPS have not been clarified. Ogose et al. reported that none of 38 patients who underwent wide excision with radiotherapyfor ASPS developed local recurrence, in contrast to 4 out of 7 patients who underwent marginal excision without radiotherapy.[9]

Multidisciplinary approach in management of these cases is preferred, complete surgical resection combined with adjuvant radiotherapy is essential for achieving long term disease free survival.

The overall survival for patients with no adjuvant treatment in ASPS is 6 months to 1 year, with radiotherapy the survival is around 3-4 years. Our patient survived for 7 years with metastatic ASPS. Long term survival is mostly as a result of adjuvant radiotherapy and chemotherapy after complete surgical excision.

Recent studies suggest the feasibility of using an antiangiogenic agent for the treatment of ASPS was demonstrated by Vistica et al. using in vivo preclinical models.[15] They observed upregulation of angiogenesis related genes in an ASPS xenograft model and demonstrated that a combination of bevacizumab (a humanized antiVEGFα monoclonal antibody) and topotecan (a topoisomerase 1 inhibitor with antiangiogenic properties) slowed the growth of the tumor by 70%.

Prognosis in these cases depends on size, presence of 17q25 cytogenetic abnormality, AJCC stage and age of patient.

Conclusion

In conclusion, ASPS though indolent but has a high propensity for metastasis. Early diagnosis and complete excision of a small primary tumor is important in the treatment of ASPS and radiotherapy plays important role in management. Antiangiogenic strategies may become a breakthrough form of management for advanced ASPS. Patients with metastatic ASPS should be enrolled in prospective clinical trials to assess the effectiveness of new treatments such as antiangiogenic therapy. Larger case series will help us understand their biological behaviour and prognosis of these patients.

References

1. J. R. Goldblum S. W. Weiss and A. L. Folpe, Enzinger and Weiss's Soft Tissue Tumors, Elsevier, Philadelphia, Pa, USA, 7th edition, 2020, chapter 33, Pages 1070-1079
2. M. Ladanyi, C. R. Antonescu, M. Drobnjak et al., "The precrystalline cytoplasmic granules of alveolar soft part sarcoma contain monocarboxylate transporter 1 and CD147," American Journal of Pathology, vol. 160, no. 4, pp. 1215–1221, 2002.
3. C. D. M. Fletcher, K. K. Unni, and F. Mertens, World Health Organization Classification of Tumours. Pathology and Genetics of Tumours of Soft Tissue and Bone, IARC Press, Lyon, France, 2002.
4. M. E. Anderson, F. J. Hornicek, M. C. Gebhardt, K. A. Raskin, and H. J. Mankin, "Alveolar soft part sarcoma: a rare and enigmatic entity," Clinical Orthopaedics and Related Research, no. 438, pp. 144–148, 2005.
5. H. E. Auerbach and J. J. Brooks, "Alveolar soft part sarcoma: a clinicopathologic and immunohistochemical study," Cancer, vol. 60, no. 1, pp. 66–73, 1987.
6. M. Casanova, A. Ferrari, G. Bisogno et al., "Alveolar soft part sarcoma in children and adolescents: A report from the soft-tissue sarcoma italian cooperative group," Annals of Oncology, vol. 11, no. 11, pp. 1445–1449, 2000.
7. A. Daigeler, C. Kuhnen, J. Hauser et al., "Alveolar soft part sarcoma: Clinicopathological findings in a series of 11 cases," World Journal of Surgical Oncology, vol. 6, article no. 71, 2008.
8. P. H. Lieberman, M. F. Brennan, M. Kimmel, R. A. Erlandson, P. Garin-Chesa, and B. Y. Flehinger, "Alveolar soft-part sarcoma. A clinico-pathologic study of half a century," Cancer, vol. 63, no. 1, pp. 1–13, 1989.
9. A. Ogose, Y. Yazawa, T. Ueda et al., "Alveolar soft part sarcoma in Japan: Multi-institutional study of 57 patients from the Japanese Musculoskeletal Oncology Group," Oncology, vol. 65, no. 1, pp. 7–13, 2003.
10. A. S. Pappo, D. M. Parham, A. Cain et al., "Alveolar soft part sarcoma in children and adolescents: clinical features and outcome of 11 patients," Medical and Pediatric Oncology, vol. 26, no. 2, pp. 81–84, 1996.

11. C. A. Portera Jr., V. Ho, S. R. Patel et al., "Alveolar soft part sarcoma: clinical course and patterns of metastasis in 70 patients treated at a single institution," Cancer, vol. 91, no. 3, pp. 585–591, 2001.
12. E. Pennacchioli, M. Fiore, P. Collini et al., "Alveolar soft part sarcoma: Clinical presentation, treatment, and outcome in a series of 33 patients at a single institution," Annals of Surgical Oncology, vol. 17, no. 12, pp. 3229–3233, 2010.
13. H. L. Evans, "Alveolar soft-part sarcoma. A study of 13 typical examples and one with a histologically atypical component," Cancer, vol. 55, no. 4, pp. 912–917, 1985.
14. S. Van Ruth, F. Van Coevorden, J. L. Peterse, and B. B. R. Kroon, "Alveolar soft part sarcoma: a report of 15 cases," European Journal of Cancer, vol. 38, no. 10, pp. 1324–1328, 2002.
15. D. T. Vistica, M. Hollingshead, S. D. Borgel et al., "Therapeutic vulnerability of an in vivo model of alveolar soft part sarcoma (ASPS) to antiangiogenic therapy," Journal of Pediatric Hematology/Oncology, vol. 31, no. 8, pp. 561–570, 2009.